HEARING LOSS IN INFANTS AND YOUNG CHILDREN:

Considerations for Pediatric Primary Care Providers

PRESENTING PATIENTS

Infants and children no older than 3 years of age with possible hearing loss initially indicated by:

- Parent concern
- Did not pass newborn hearing screening
- One or more risk factors for hearing loss, regardless of screening result (See Risk Indicators section)
- Audiologic or other test results suggesting hearing loss in one or both ears

No child is too young to test: Delay in obtaining intervention services can compromise language development and reduce communication and school performance. **Remember 1-3-6:**

► **By 1 month of age** | Obtain hearing screen no later than 1 month of age if no hearing screening has been done.

► **By 3 months of age** | Refer immediately to pediatric audiologist **if child did not pass hearing screening and audiologic evaluation[1] has not been completed**. Hearing status should be confirmed by audiologic evaluation no later than 3 months of age.

► **By 6 months of age** | Verify entry into Early Intervention[2] services no later than 6 months of age **if hearing loss is confirmed**.

If child passes hearing screening but displays risk factors, including caregiver concern: Assess risk factor(s) and plan audiologic monitoring with caretakers. Late-onset hearing loss can occur at any time after birth despite a passed hearing screen at birth[3].

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KEY PRACTICE POINTS

- Refer all infants who do not pass hearing screening for diagnostic audiologic evaluation.
- Once any degree of hearing loss is diagnosed (including "transient" hearing loss associated with cleft palate), referral to Early Intervention must be made within 7 working days of confirmation of hearing loss.
- Ongoing care for all hearing losses ("transient" and permanent) by an audiologist is essential, in addition to medical care.
- For children who passed their birth-screen but are at risk for hearing loss: ensure that a diagnostic audiologic evaluation is completed at least once by 24 to 30 months of age[4].
- Regardless of previous hearing screening outcomes, all infants with or without risk factors should receive continued tracking of auditory skills, language milestones, and middle ear status during well-child visits in the medical home.

RECOMMENDATIONS FOR CLINIC VISIT

HISTORY

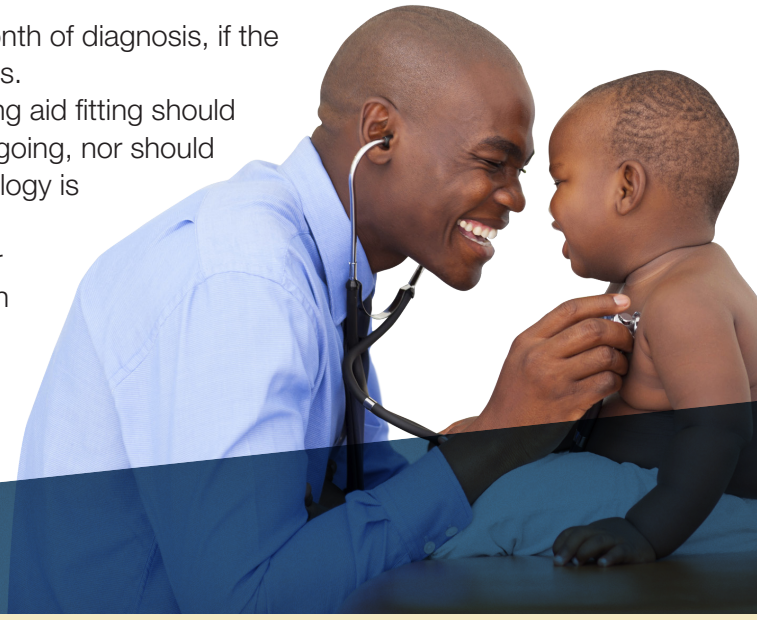
- Review all medical records and case history information for risk factors associated with congenital hearing loss and/or late-onset or progressive hearing loss (See below Risk Indicator section).
- Review audiologic record to assess need for further audiologic testing.
- Elicit whether there is parental concern about the child's hearing.
- Pay special attention to dysmorphic features, and presence of ear pits or branchial cysts.
- If hearing loss etiology remains undetermined consider referring family to a genetic specialist to determine possible genetic etiology.

PHYSICAL EXAM & REFERRAL

- Every infant with confirmed hearing loss should be evaluated by an otolaryngologist who has knowledge of pediatric hearing loss and have at least one visual acuity examination by an ophthalmologist who is experienced in evaluating infants.
- Verify with parents if referral to Early Intervention has been made[2].
- Also ask parents if referral to parent support has been done. Many state Early Hearing Detection & Intervention (EHDI) programs have parent support resources http://www.infanthearing.org/states/index.html

TREATMENT

- Hearing aid fitting should take place within one month of diagnosis, if the child's parents/caregivers have chosen hearing aids.
- Medical clearance by an otolaryngologist for hearing aid fitting should not be delayed while other medical exams are on-going, nor should such clearance be delayed until a diagnosis of etiology is made.
- Hearing aid fitting when there is chronic middle ear effusion should be considered if other options such as myringotomy or tube insertion are not possible, especially when associated with cleft palate and Down Syndrome.



RISK INDICATORS

Risk indicators[5] associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood: Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.
2. Family history§ of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: Extracorporeal Membrane Oxygenation (ECMO)§, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion. *Any child who has been in the neonatal intensive care unit (NICU) for 5 or more days requires an Automated Auditory Brainstem Response (AABR) by an audiologist*
4. In utero infections, such as Cytomegalovirus (CMV)§, herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. Neurodegenerative disorders§, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss§, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
11. Chemotherapy§.

WEBSITES

TO LOCATE:

- **Pediatric audiology services -** Early Hearing Detection & Intervention- Pediatric Audiology Links to Services (EHDI-PALS) http://ehdipals.org
- **State Early Hearing Detection and Intervention Programs -** National Center for Hearing Assessment and Management http://www.infanthearing.org/states

OTHER RESOURCES

- American Academy of Audiology www.audiology.org
- American Academy of Family Physicians Patient-Centered Medical Home Checklist http://www.aafp.org/practice-management.html
- American Academy of Otolaryngology Head and Neck Surgery www.entnet.org
- American Academy of Pediatrics Newborn and Infant Hearing Screening Activities http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx
- American Speech-Language-Hearing Association (ASHA) www.asha.org
- CDC Early Hearing Detection and Intervention (EHDI) www.cdc.gov/ncbddd/ehdi/
- *Joint Committee on Infant Hearing (JCIH) www.jcih.org

FOOTNOTES

1. A complete audiologic evaluation for infants consists of (may vary depending on age, circumstances, diagnosis):

- Acoustic immittance (tympanometry and acoustic reflex thresholds {high-frequency probe tone for infants under 6 months of age})
- Otoacoustic emissions (Distortion Product or Transient)
- Auditory Brainstem Response (Air- and bone-conduction) for specific frequencies (in particular, 500 Hz and 2000 Hz, others as time permits). ABR protocols to assess possible Auditory Neuropathy when indicated.
- Auditory Steady-State Response as a secondary measure (after tone-burst ABR).
- Behavioral Audiometry at age- and developmentally-appropriate stages.

2. Early Intervention according to Part C of IDEA 2004 is the process of providing services, education and support to young children who are deemed to have

- an established condition,
- a diagnosed physical or mental condition (with a high probability of resulting in a developmental delay),
- an existing delay or
- a child who is at-risk of developing a delay or
- special need that may affect their development or impede their education.
  http://ectacenter.org/partc/partc.asp
  http://pediatrics.aappublications.org/content/early/2013/03/18/peds.2013-0008.citation

3. Newborn Hearing Screening: Lost to Documented Follow up Considerations for the Medical Home, AAP News Aug. 2013. http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/pehdic/pages/early-hearing-detection-and-intervention.aspx

4. Guidelines for Rescreening in the Medical Home Following a "Do Not Pass" Newborn Hearing Screening, AAP http://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Documents/Rescreening_Guidelines.PDF

5. Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs: Joint Committee on Infant Hearing. Pediatrics Vol. 120 No. 4 October 1, 2007 pp. 898-921 (doi: 10.1542/peds.2007-2333) http://pediatrics.aappublications.org/content/120/4/898.full?ijkey=oj9BAleq21OlA&keytype=ref&siteid=aapjournals